



## Congenital Heart Disease Cases

Sabrina Phillips, MD FACC FASE  
Mayo Clinic Congenital Heart Disease Center

©2013 MAYO CLINIC

No Disclosures



©2013 MAYO CLINIC

## CASE 1



© 2013 Mayo Clinic. All rights reserved.

### 63 year old Woman

- Healthy throughout life except:
  - Rheumatic fever at age 7
  - Palpitations for several years
- 2 months ago noticed increasing shortness of breath



© 2013 Mayo Clinic. All rights reserved.

## Recent Hospitalization

- Anasarca - responded to diuresis
- Atrial fibrillation – treated with rate control and anti-coagulation



© 2013 Mayo Clinic. All rights reserved.

## Physical Examination

- Neck: JVP is moderately elevated with prominent V wave
- Heart:
  - 2+ RV; Normal LV impulse
  - Irreg RR; soft S<sub>1</sub>; single S<sub>2</sub>; S<sub>3</sub> present
  - III/VI systolic murmur
  - I/IV diastolic murmur at L sternal border
- Abdomen: Liver is pulsatile 4cm below costal margin
- Ext: Trace edema



© 2013 Mayo Clinic. All rights reserved.

## Medications

- Aspirin 81 mg QD
- Furosemide 40 mg QD
- Lanoxin 250 mcg tablet QD
- Metoclopramide 10 mg TID
- Metoprolol Tartrate 50 mg BID
- Potassium Chloride 20 mEq QD
- Warfarin 3 mg QD



© 2013 ARMED, J. 000000

## Exercise Stress Test

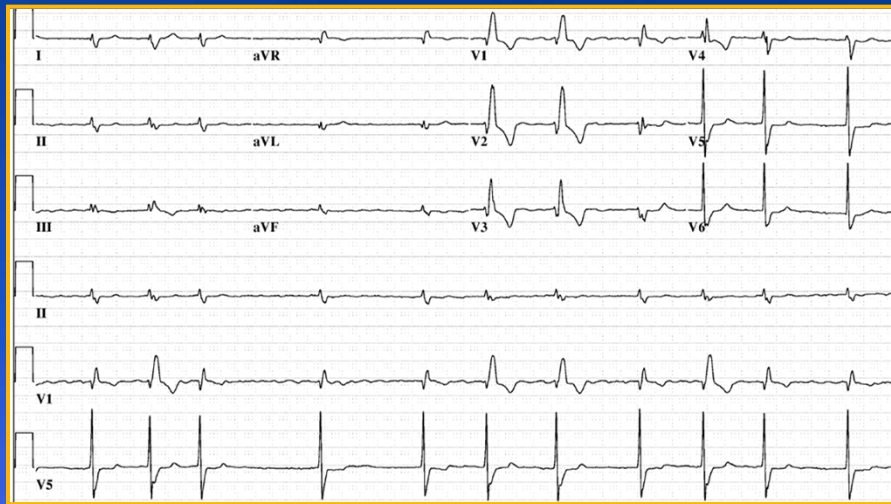
- Exercise Time: 3.8 minutes
- FAC: 48%
- Peak  $\text{VO}_2 = 13.4 \text{ mL/kg/min}$  (59%)



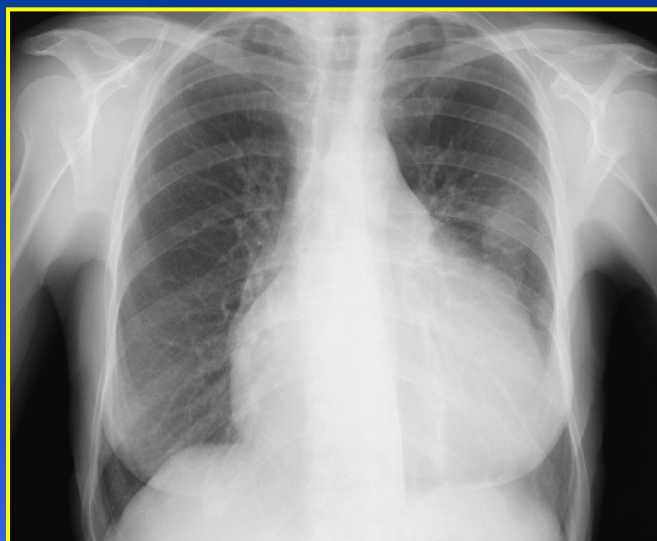
© 2013 ARMED, J. 000000

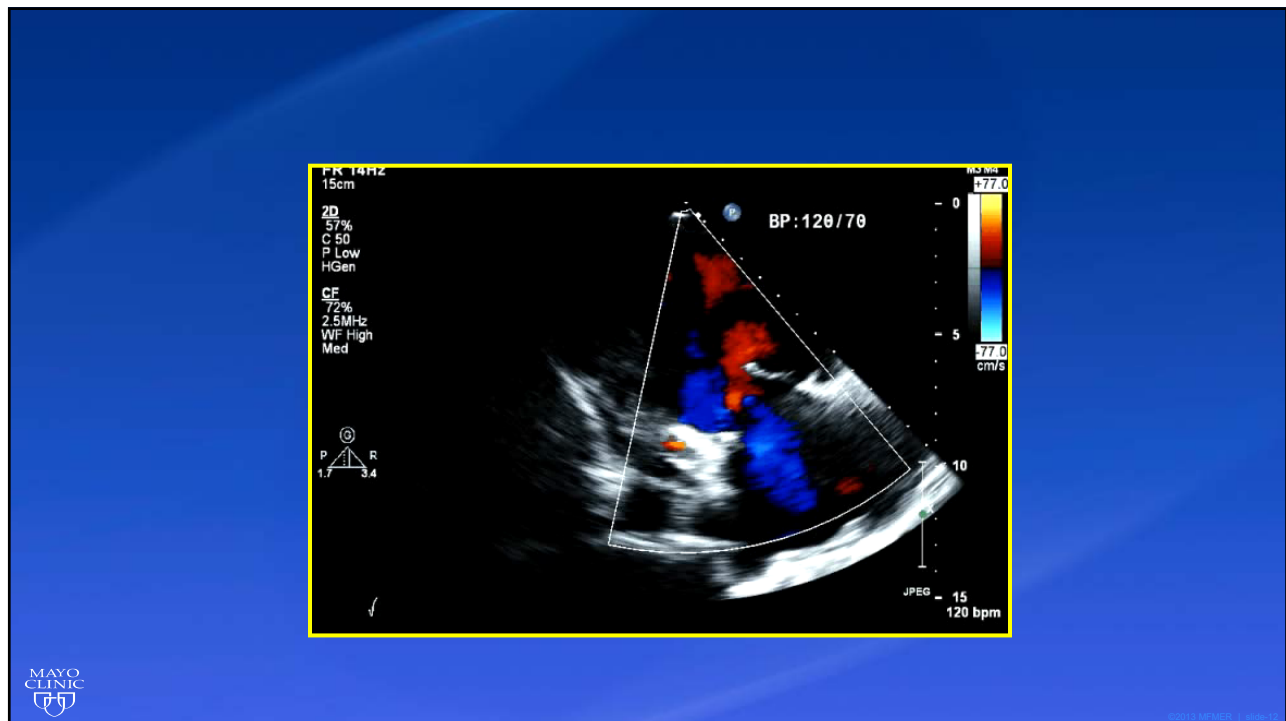
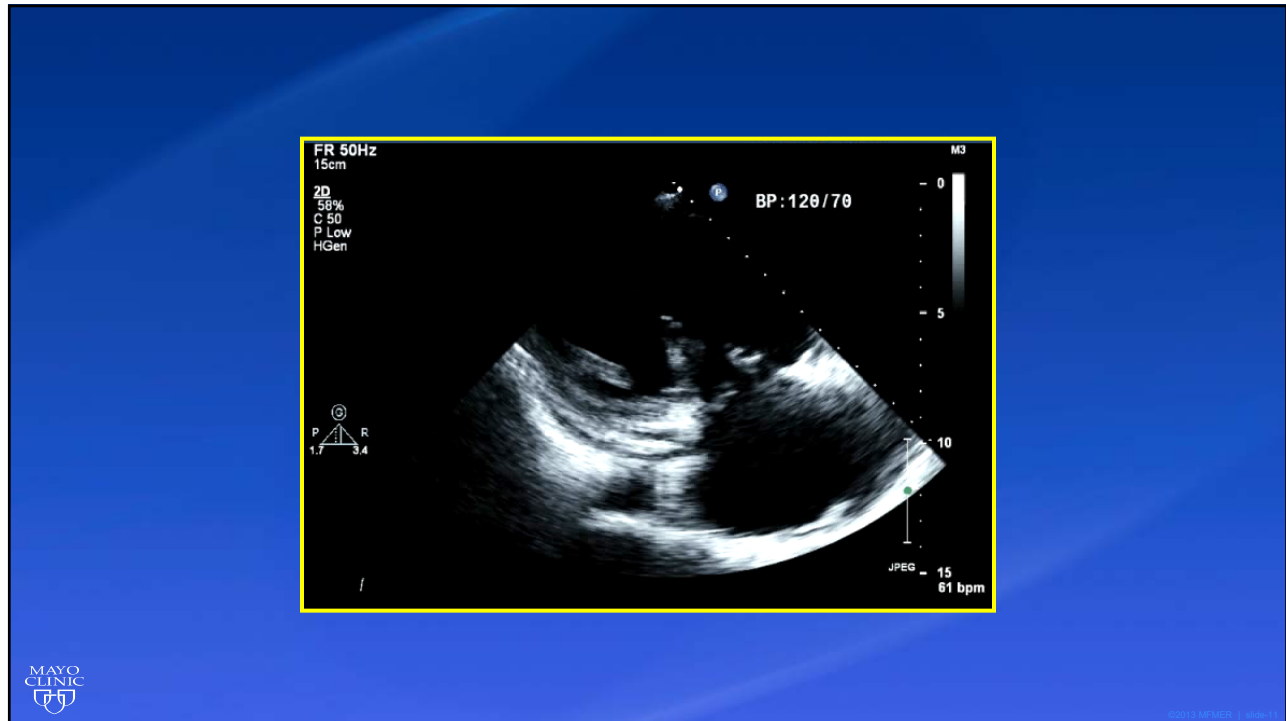


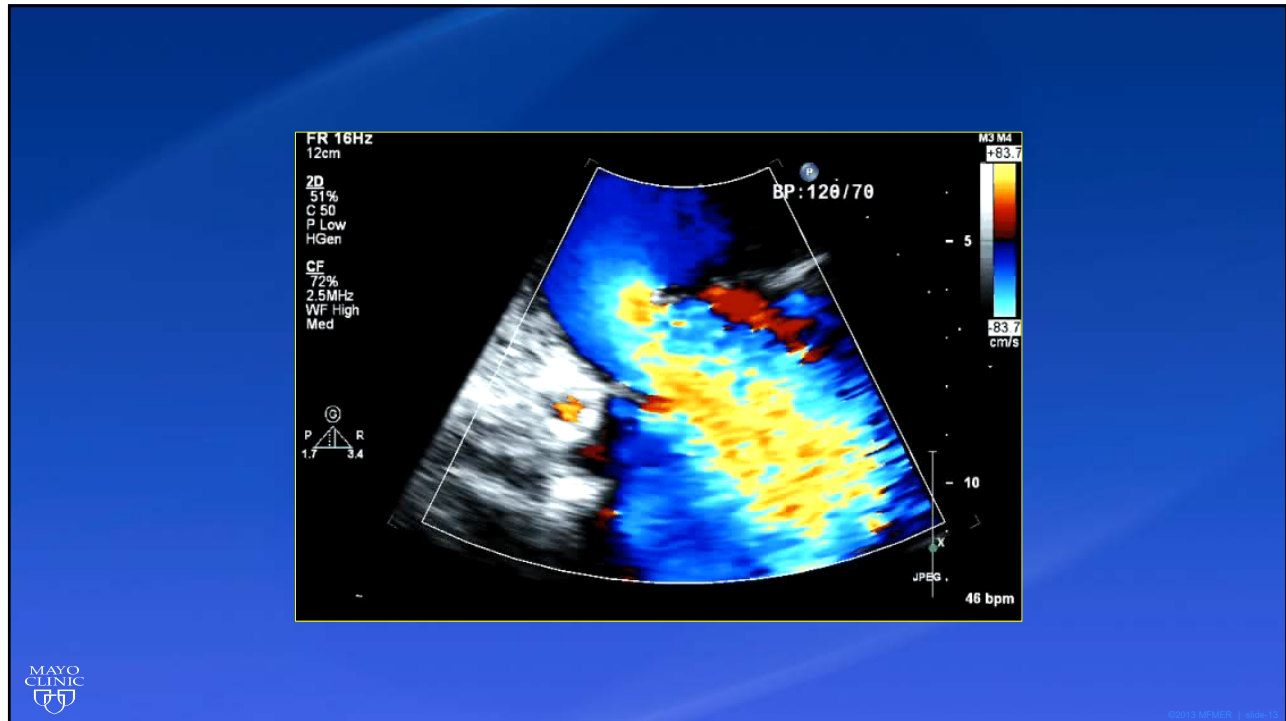
## ECG

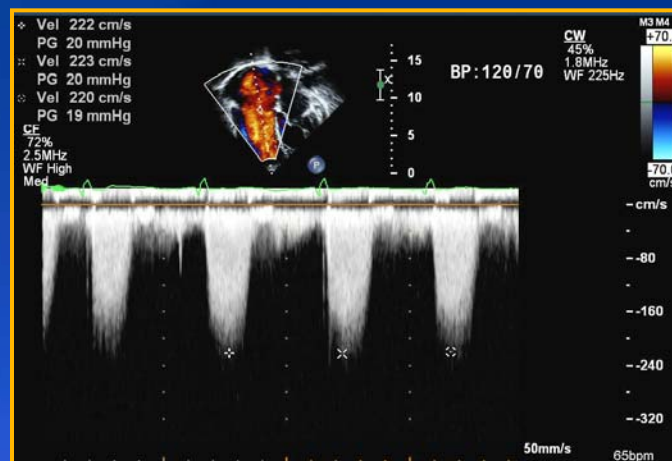
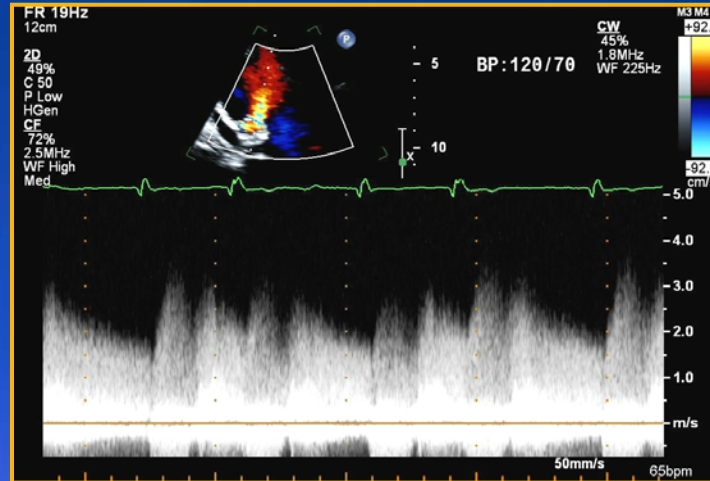


## Chest X-Ray

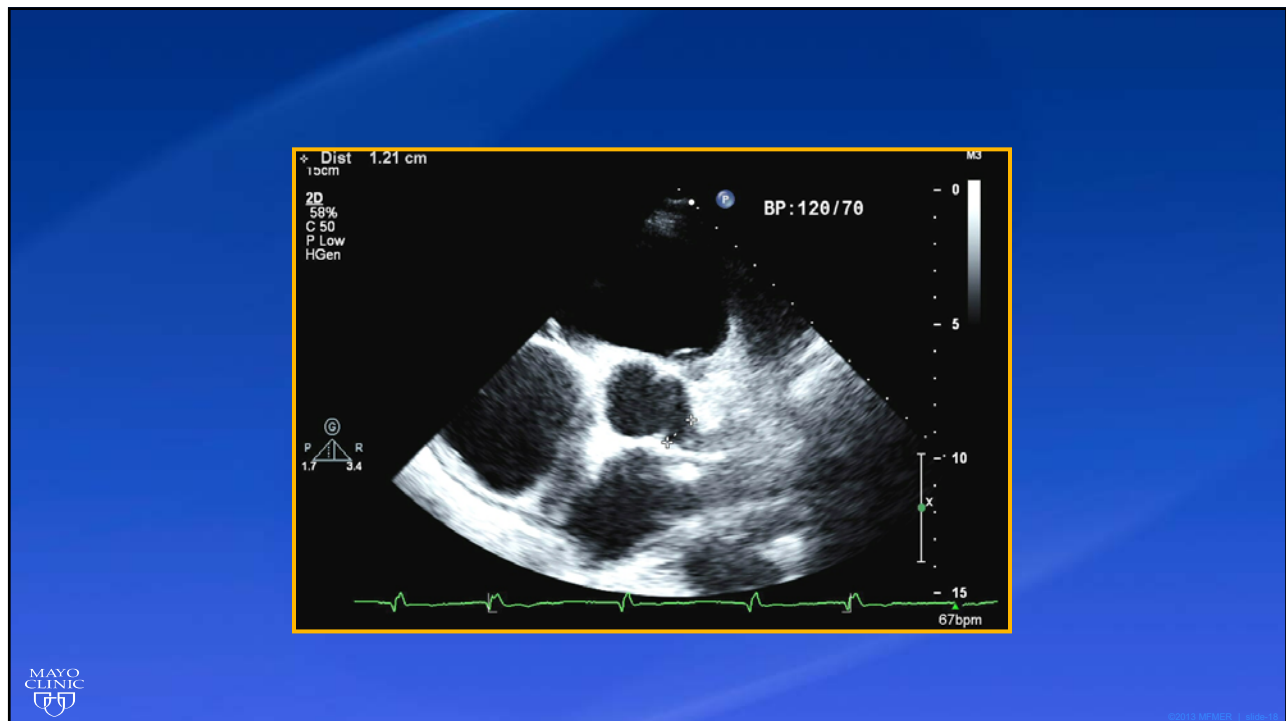
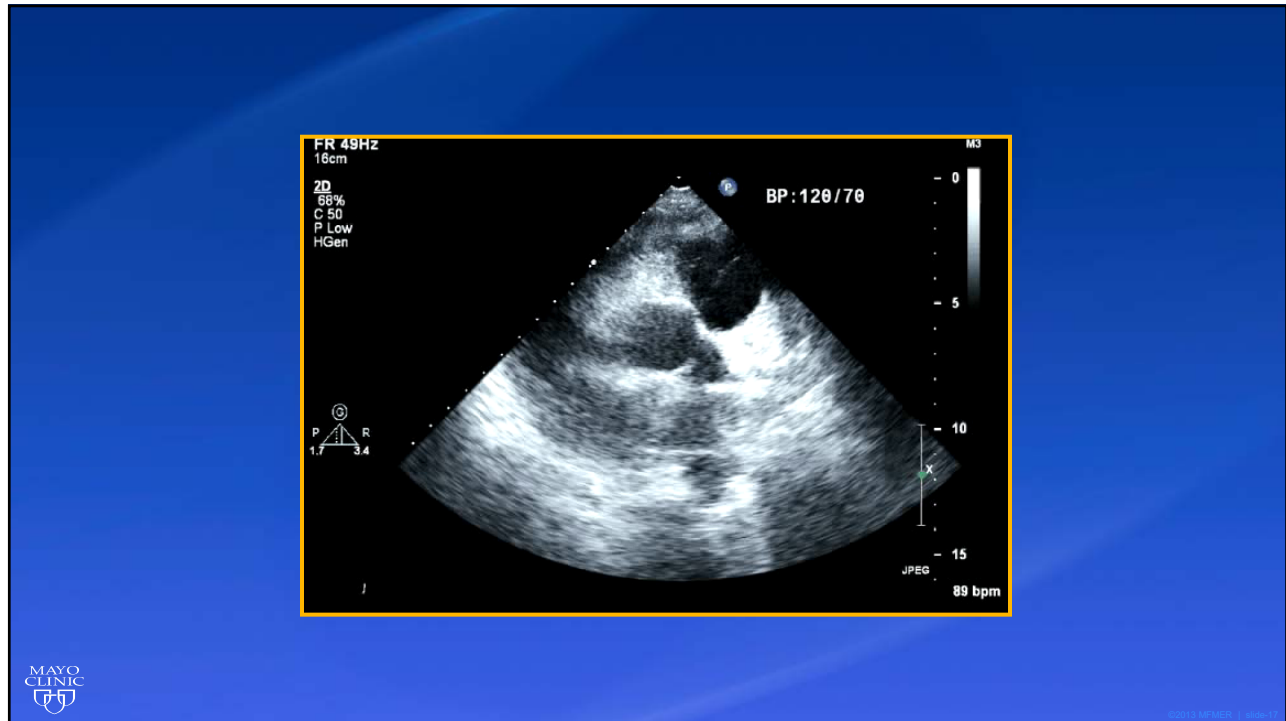


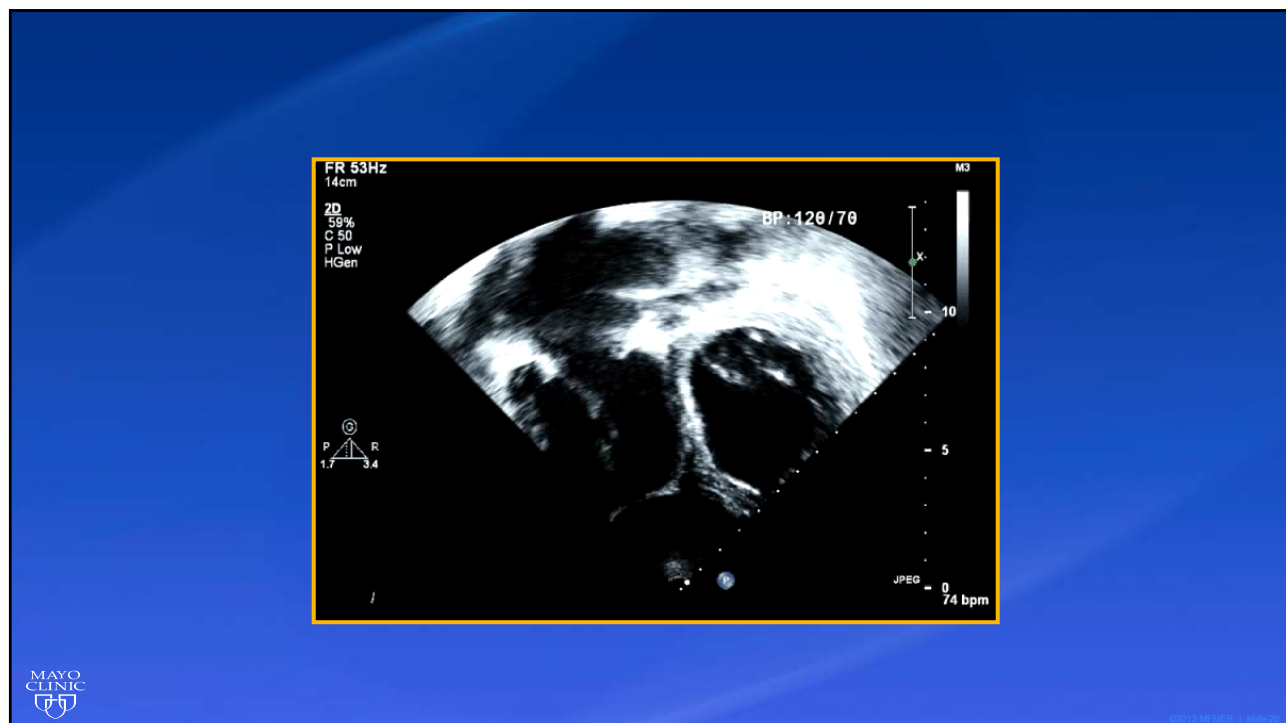


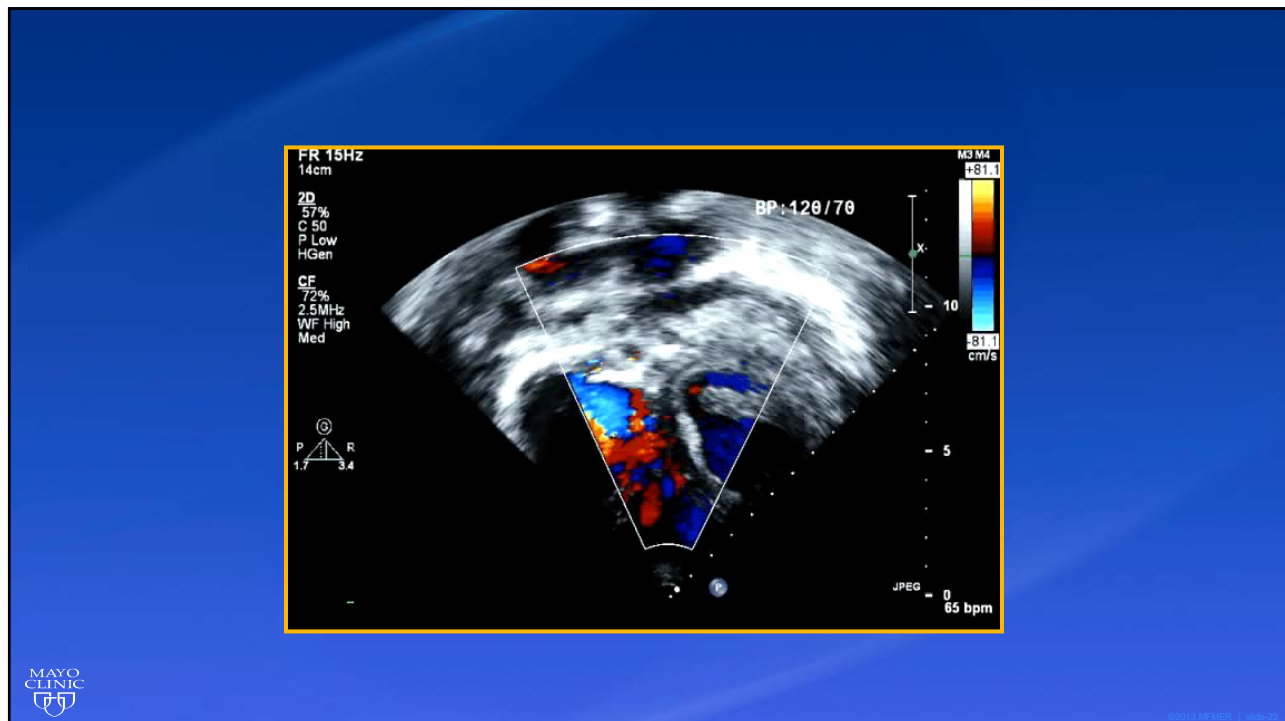
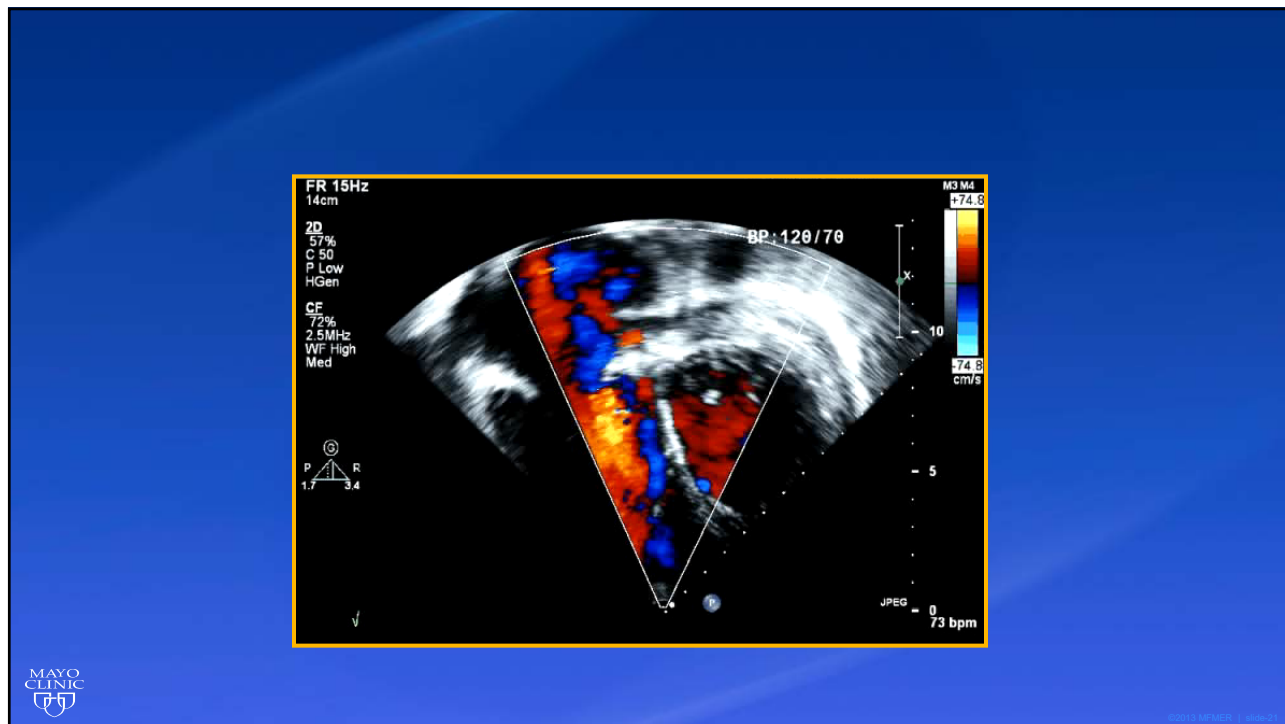




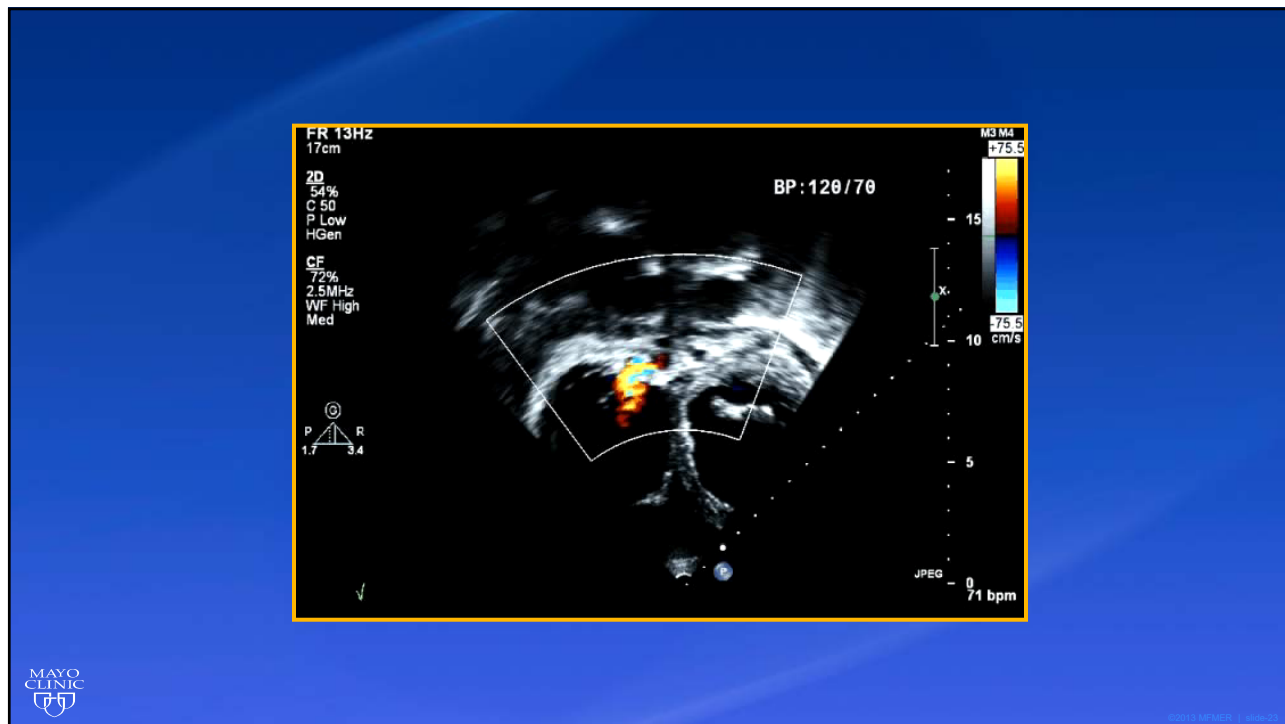
RVSP = 29 mmHg





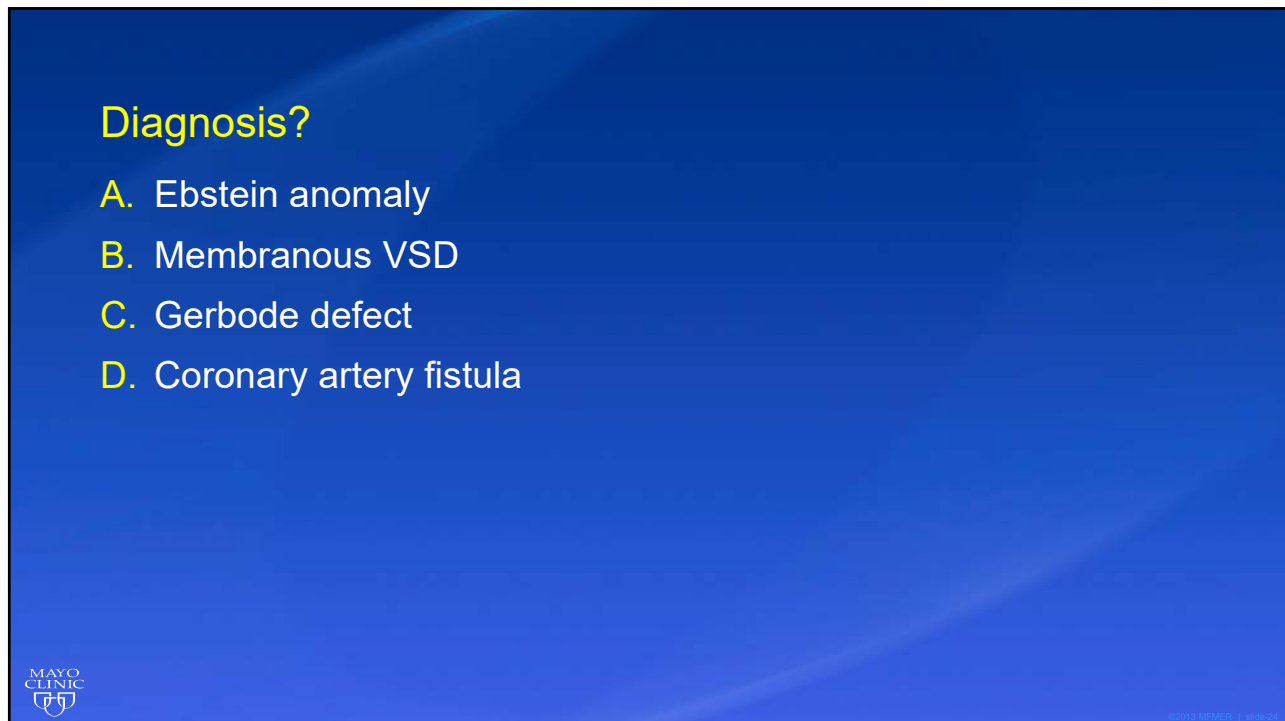






## Diagnosis?

- A. Ebstein anomaly
- B. Membranous VSD
- C. Gerbode defect
- D. Coronary artery fistula





## Diagnosis?

- A. Ebstein anomaly
- B. Membranous VSD
- C. Gerbode defect
- D. **Coronary artery fistula**



©2013 Mayo Clinic. All rights reserved.

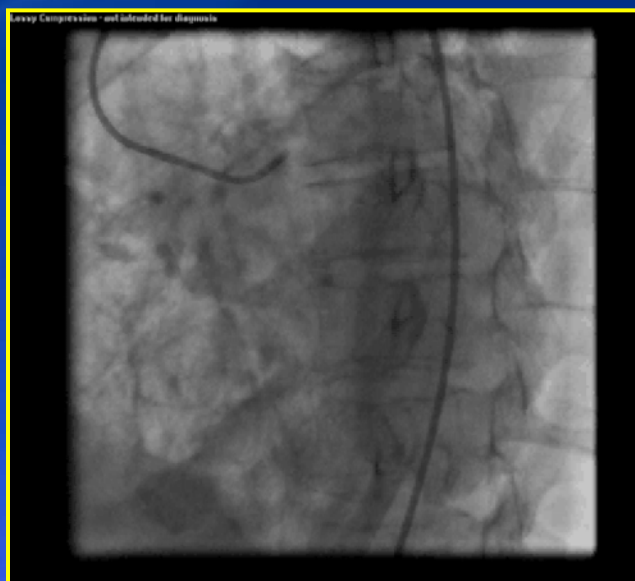
## Echo Results

- Enlarged left coronary system
  - Circumflex to RV fistula
- Flail Tricuspid valve leaflet with severe TR
- Severe RV enlargement with mildly decreased function; RVSP = 29 mmHg
- Severe RA enlargement
- LV EF = 58%



©2013 Mayo Clinic. All rights reserved.

## Catheterization

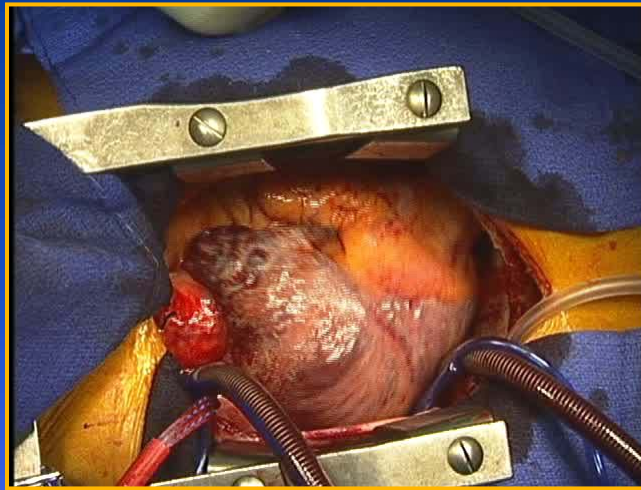




## What Next?

- A. Catheter intervention
- B. Surgical consultation
- C. More testing

## Surgical Intervention



©2013 Mayo Clinic. All rights reserved.

## Surgery

- Repair of coronary artery fistula with 2 layer primary closure in the RV
- 31-mm Epic porcine tricuspid valve replacement
- PFO closure
- Radiofrequency maze procedure
- Uncomplicated hospital course



©2013 Mayo Clinic. All rights reserved.

## Case 2

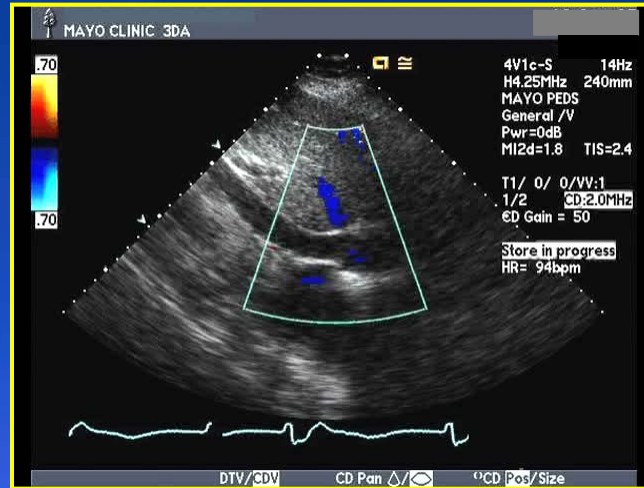


### 26 Year old Woman

- Diagnosed with tetralogy of Fallot in the neonatal period
- Operative repair at age 2
- Sporadic follow up after age 12
- Presented with progressive decline in stamina
- Elsewhere underwent mitral valve replacement with a tissue prosthesis for “mitral valve prolapse” and regurgitation
- Presents 4 weeks after surgery with intractable pleural effusions, fatigue and high grade AV block



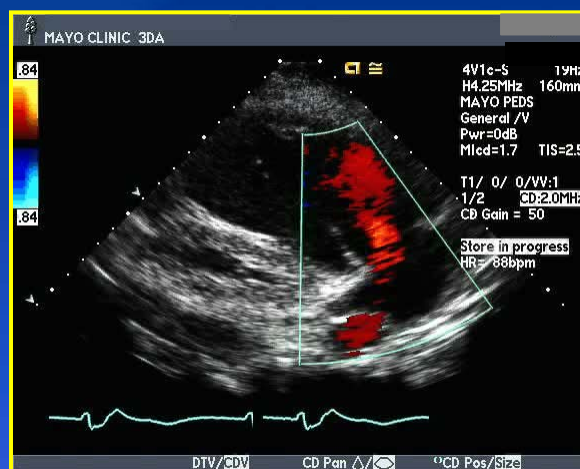
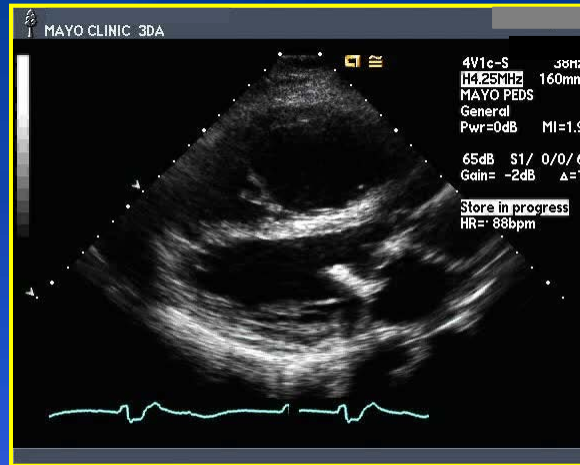
# Echocardiogram

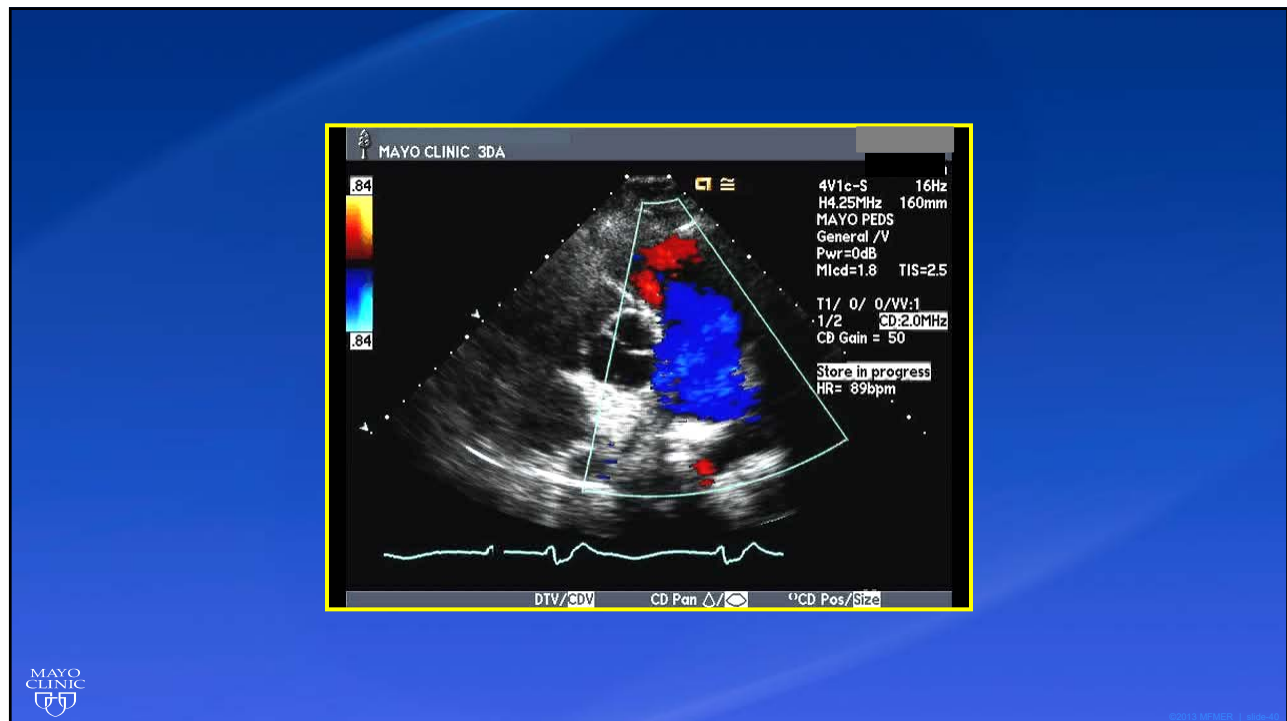
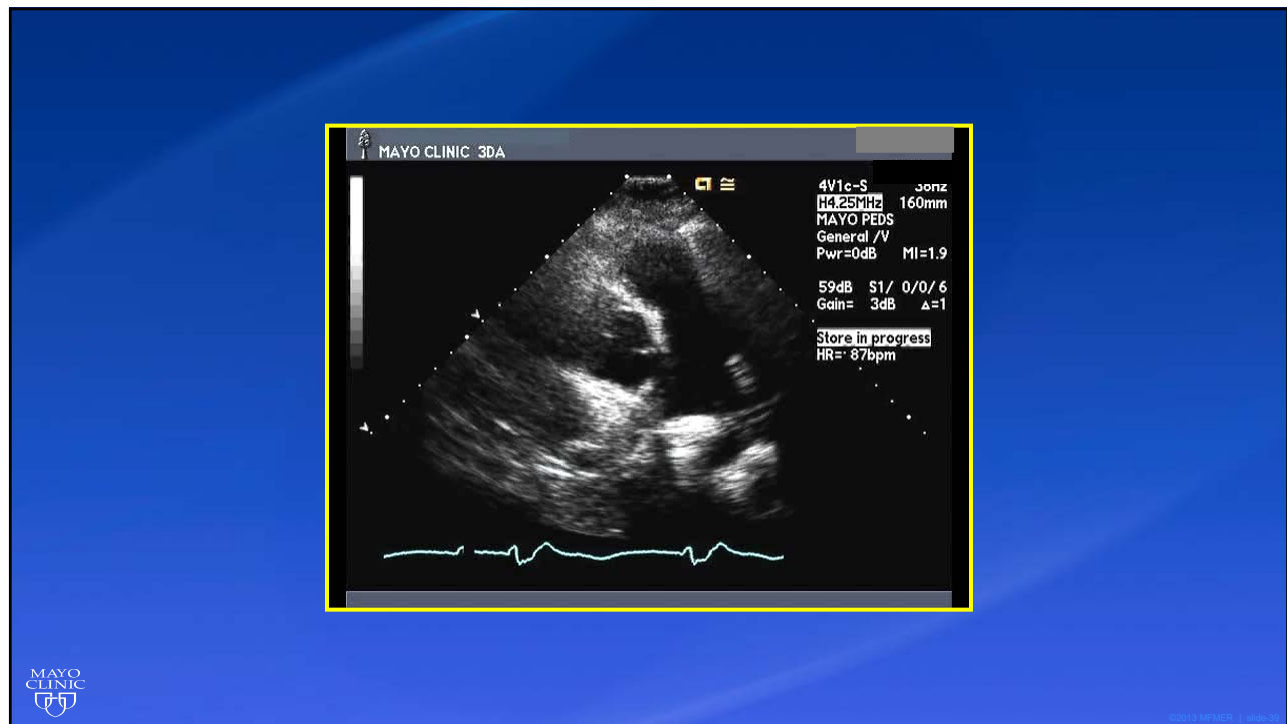


©2013 MEDICAL



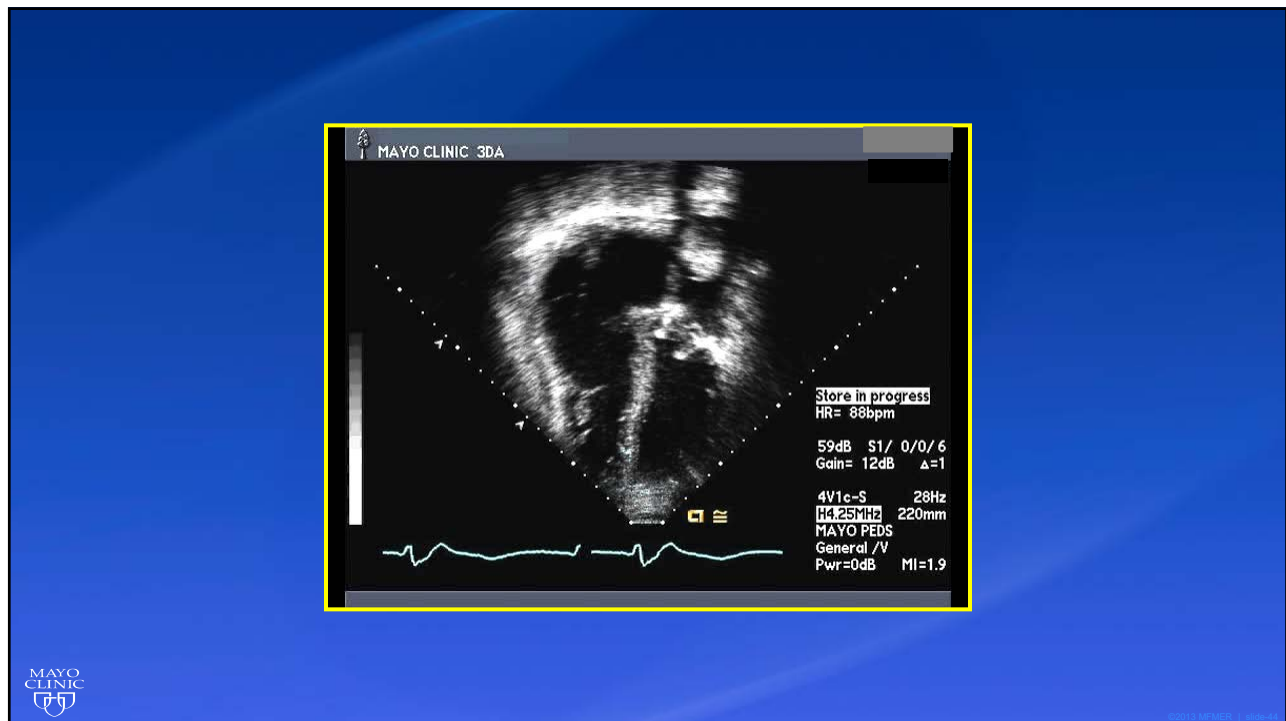
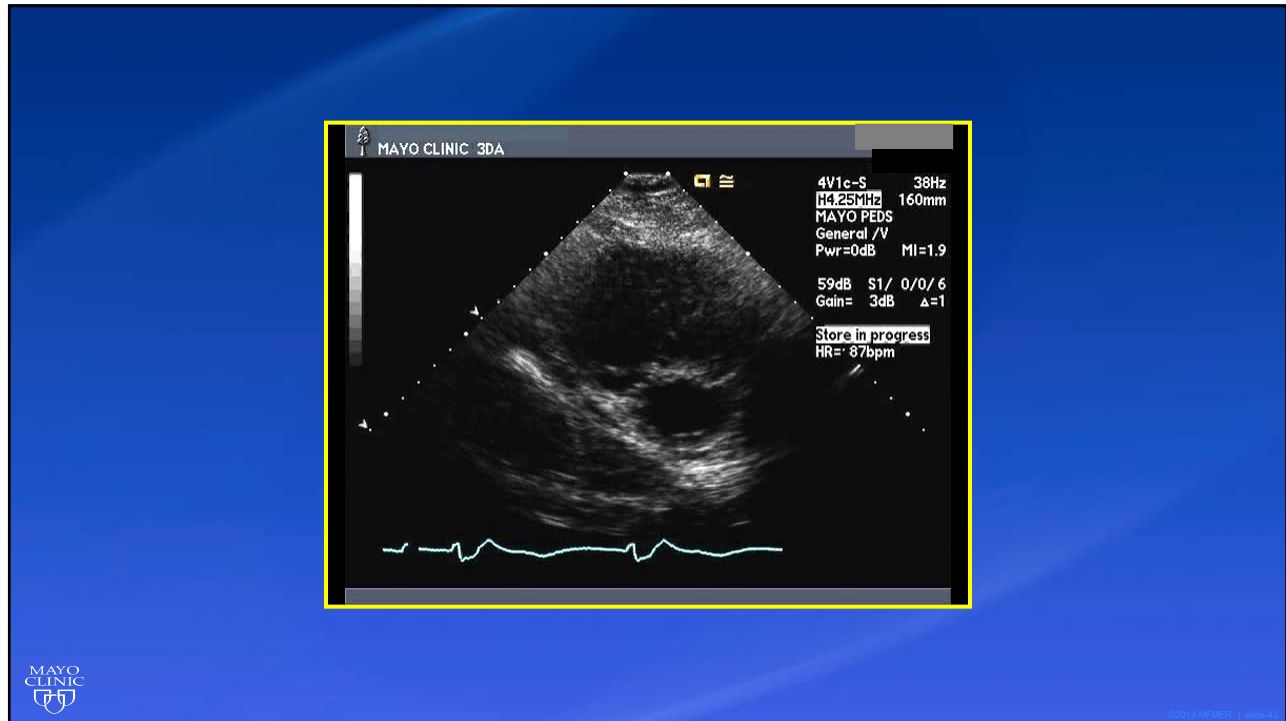
©2013 MEDICAL

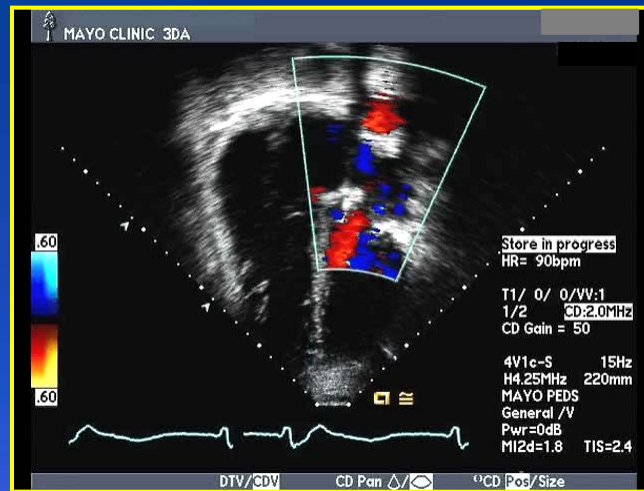




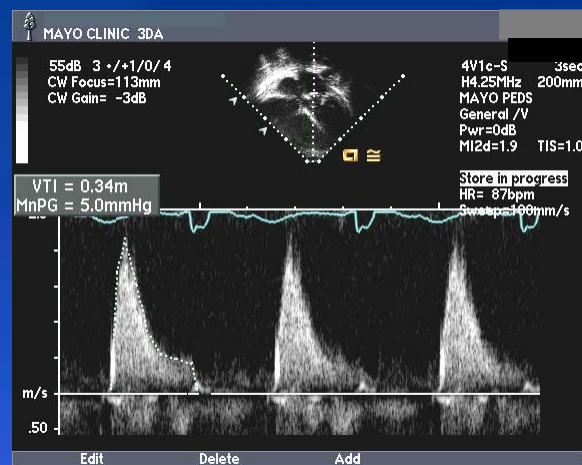




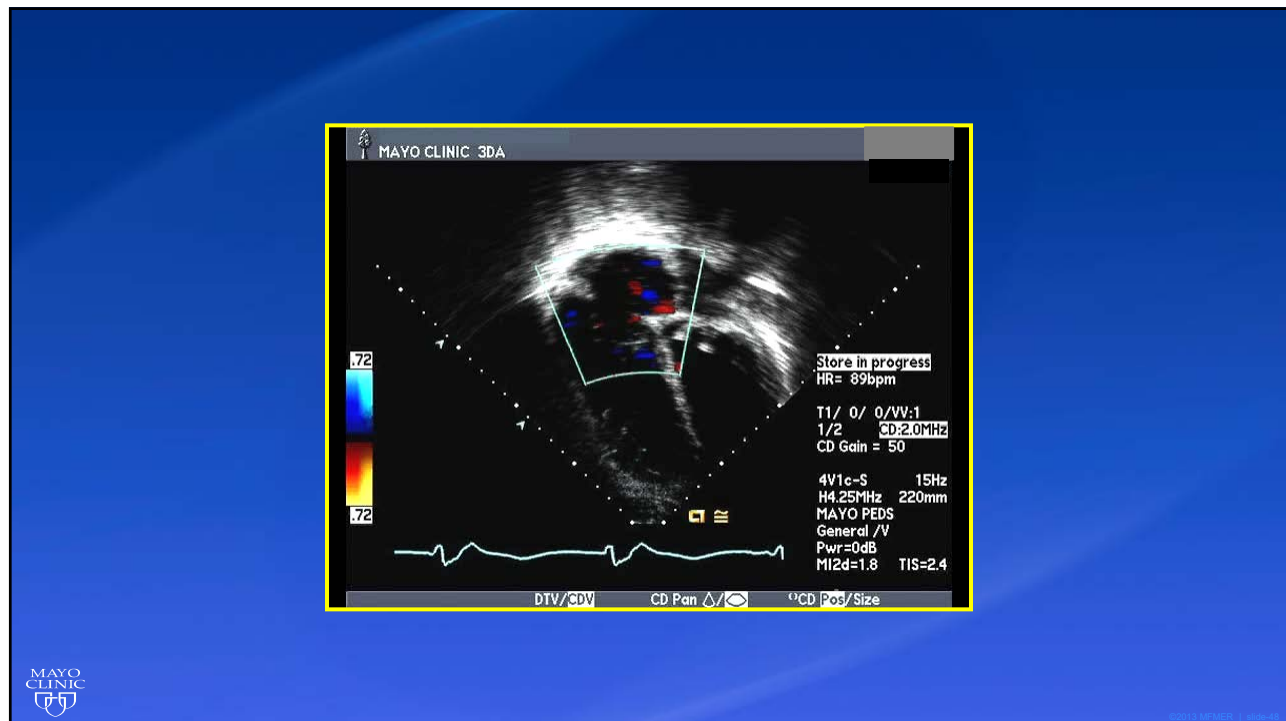
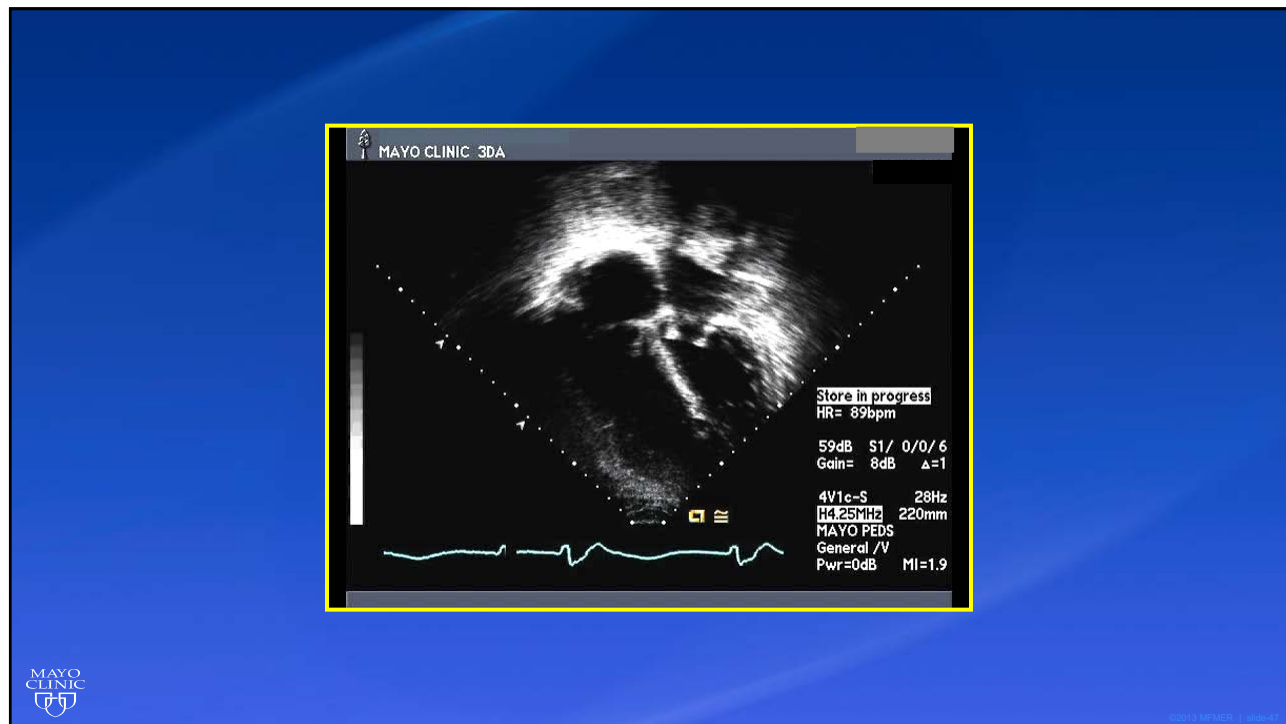


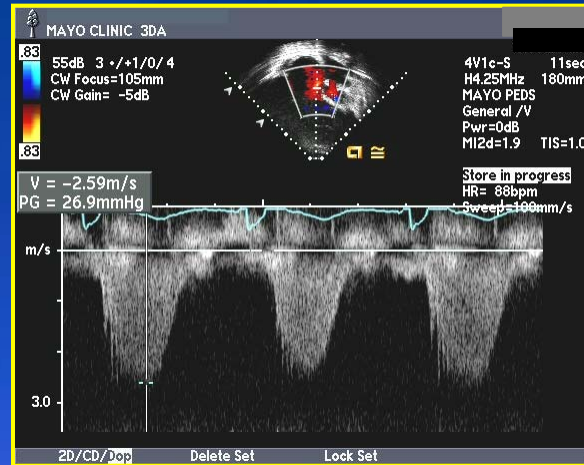


©2013 MEDICAL ultrasound



©2013 MEDICAL ultrasound





©2013 MEDICAL

## Diagnosis?

- A. Constriction
- B. Severe tricuspid valve regurgitation
- C. Severe pulmonary valve regurgitation
- D. Pulmonary hypertension



©2013 MEDICAL

## Diagnosis?

- A. Constriction
- B. Severe tricuspid valve regurgitation
- C. Severe pulmonary valve regurgitation
- D. Pulmonary hypertension



©2013 Mayo Clinic. All rights reserved.

## ECHO Report

- Severe RV enlargement, moderate-severe decrease in function, RVSP 39 mmHg.
- Severe (free) pulmonary regurgitation
- LV EF 30% - 35%
- Abnormal hepatic vein Doppler related to junction rhythm
- Normal mitral tissue prosthesis

Patient medically optimized then referred for PVR



©2013 Mayo Clinic. All rights reserved.

## Post-Operative Hemodynamic Concerns

- Pulmonary valve regurgitation
- Residual/recurrent RVOT obstruction
- Residual VSD
- Tricuspid valve regurgitation
- Aortic root enlargement +/- aortic valve regurgitation



©2013 Mayo Clinic. All rights reserved.

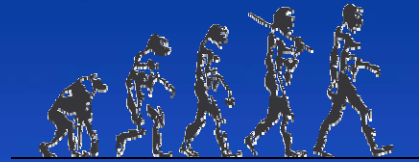
## Consequences of Pulmonary Valve Regurgitation

- Exercise intolerance
- Right ventricular dilatation
- Right ventricular dysfunction
- Increased risk of ventricular tachycardia
- Increased risk of atrial arrhythmia
- Left ventricular dysfunction



©2013 Mayo Clinic. All rights reserved.

## Evolution of PVR Timing



Pulmonary  
Valve Not  
Important



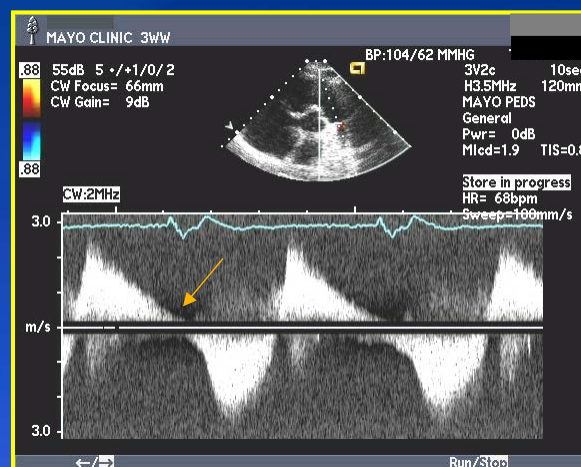
Pulmonary  
valve should be  
replaced for  
right heart  
failure  
symptoms



Pulmonary valve should  
be replaced to prevent  
right ventricular  
dysfunction



©2013 Mayo Clinic. All rights reserved.



©2013 Mayo Clinic. All rights reserved.



## Case 3



©2013 Mayo Clinic. All rights reserved.

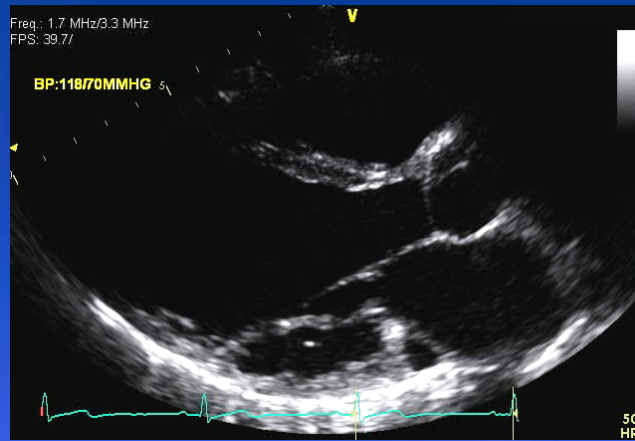
### 24 year old

- Denies complaints
- Echo obtained after murmur was heard on exam for assessment to be kidney donor

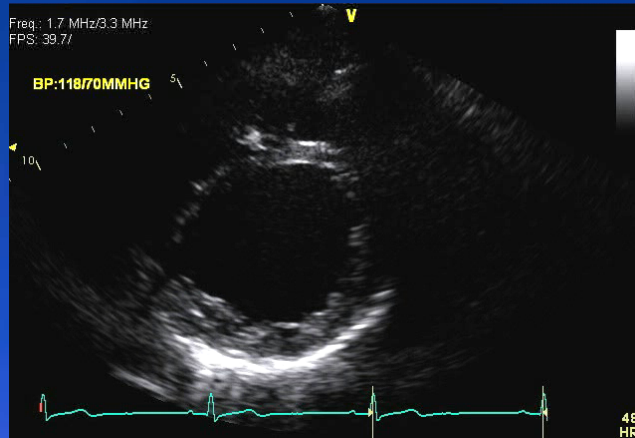


©2013 Mayo Clinic. All rights reserved.

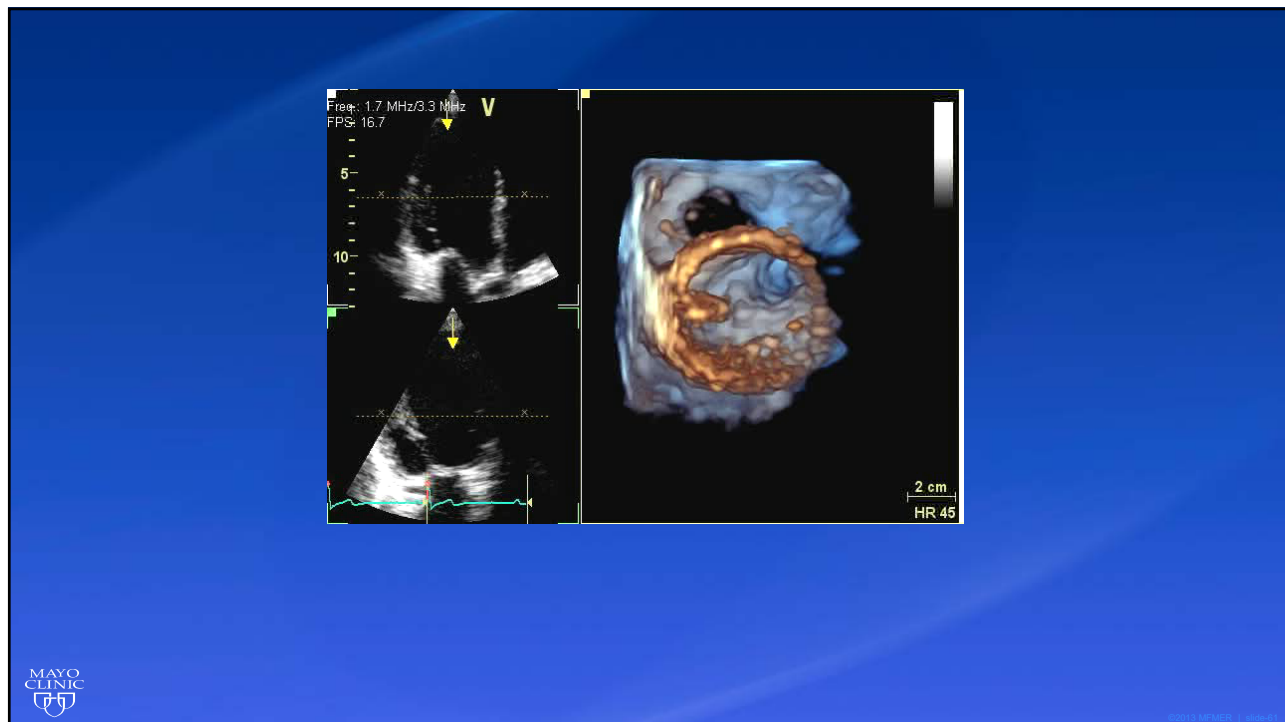
## 24 year Old : Wants to be kidney donor



©2013 Mayo Clinic



©2013 Mayo Clinic



## Diagnosis?

- A. Parachute mitral valve
- B. Cleft mitral valve
- C. Supravalvular mitral ring
- D. Rheumatic mitral stenosis

## Diagnosis?

- A. Parachute mitral valve
- B. Cleft mitral valve
- C. Supravalvular mitral ring
- D. Rheumatic mitral stenosis



©2013 Mayo Clinic. All rights reserved.

## Parachute Mitral Valve

- Abnormal compaction of ventricular trabecular myocardium and abnormal delamination of the trabecular ridge
- Unifocal attachment of the mitral valve cordae to a single/fused papillary muscle
- Papillary muscle usually centrally placed
- Often associated with other left heart abnormalities
- Mitral stenosis most common hemodynamic consequence



©2013 Mayo Clinic. All rights reserved.

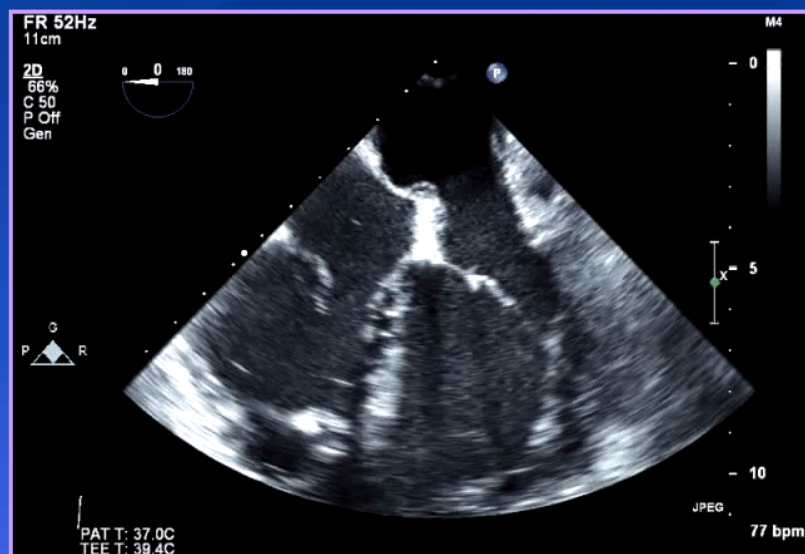
## Mitral Ring

- Supramitral ring: fibrous membrane just above the mitral annulus. Does not adhere to the leaflets. Subvalvular apparatus normal
- Intramitral ring: thin membrane within the funnel created by the valve leaflets. Always combined with an abnormal subvalvular apparatus
- Results in stenosis

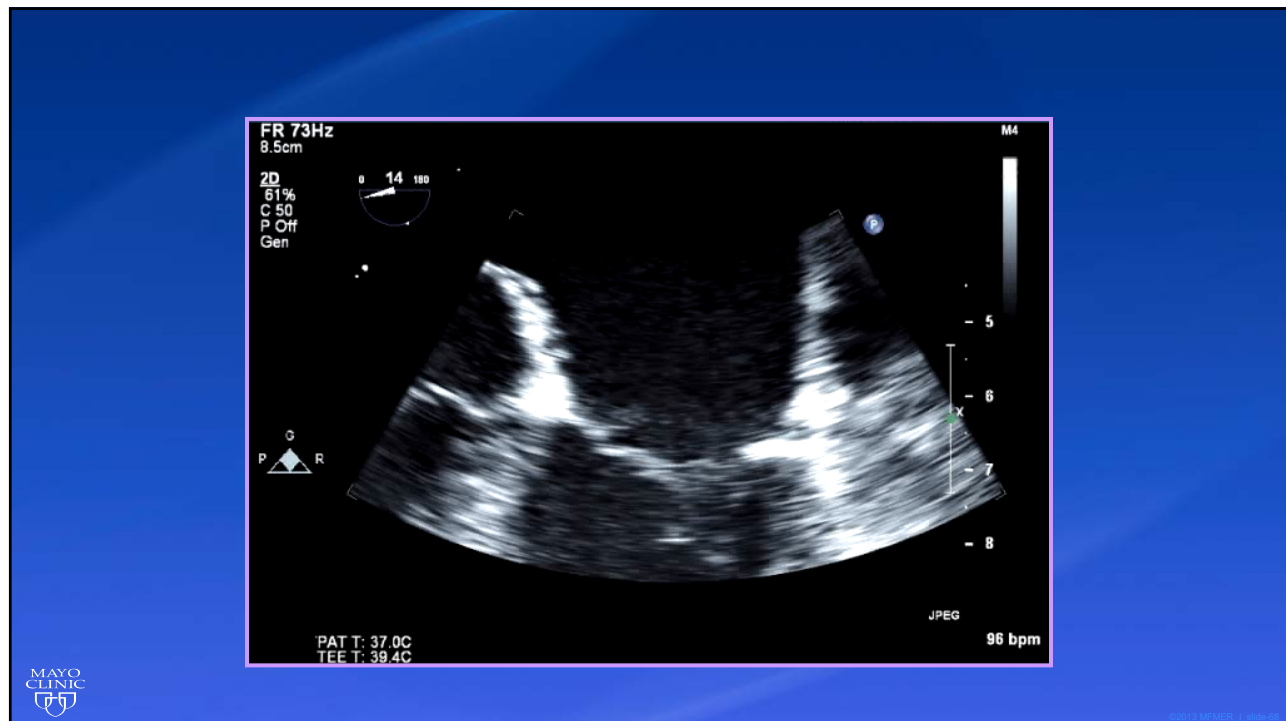
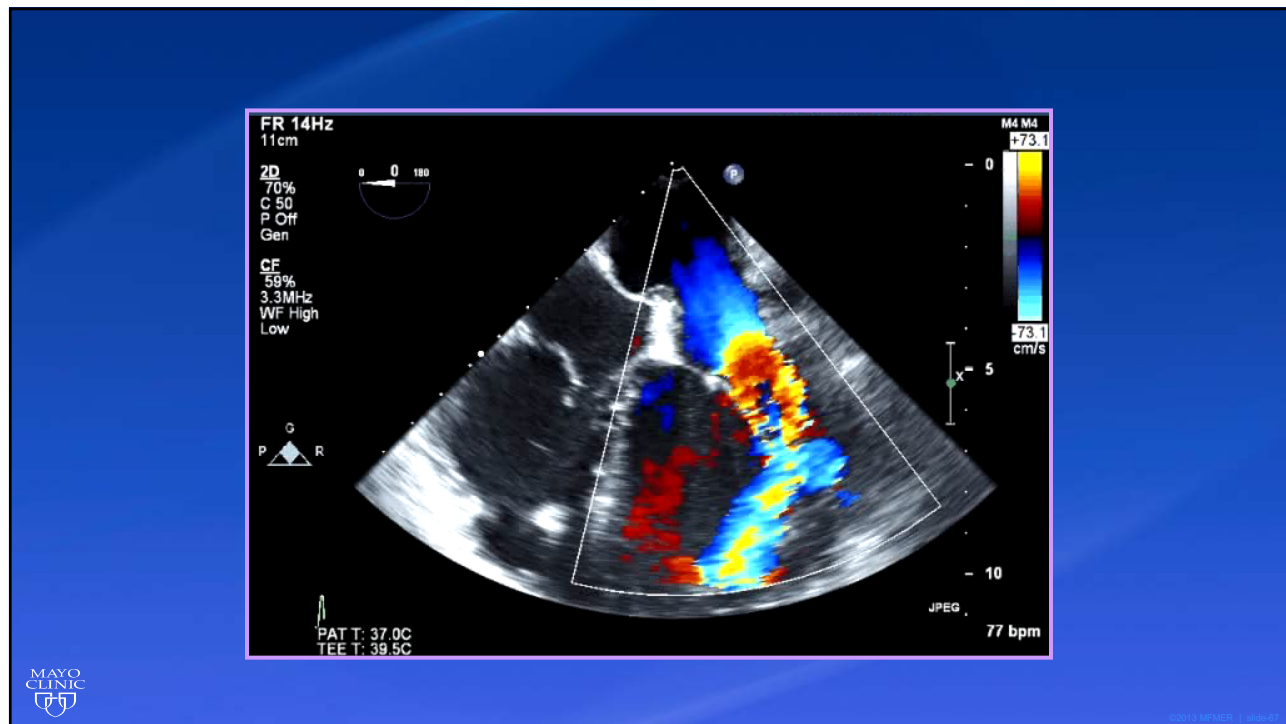


©2013 Mayo Clinic

## Supramitral Ring



©2013 Mayo Clinic





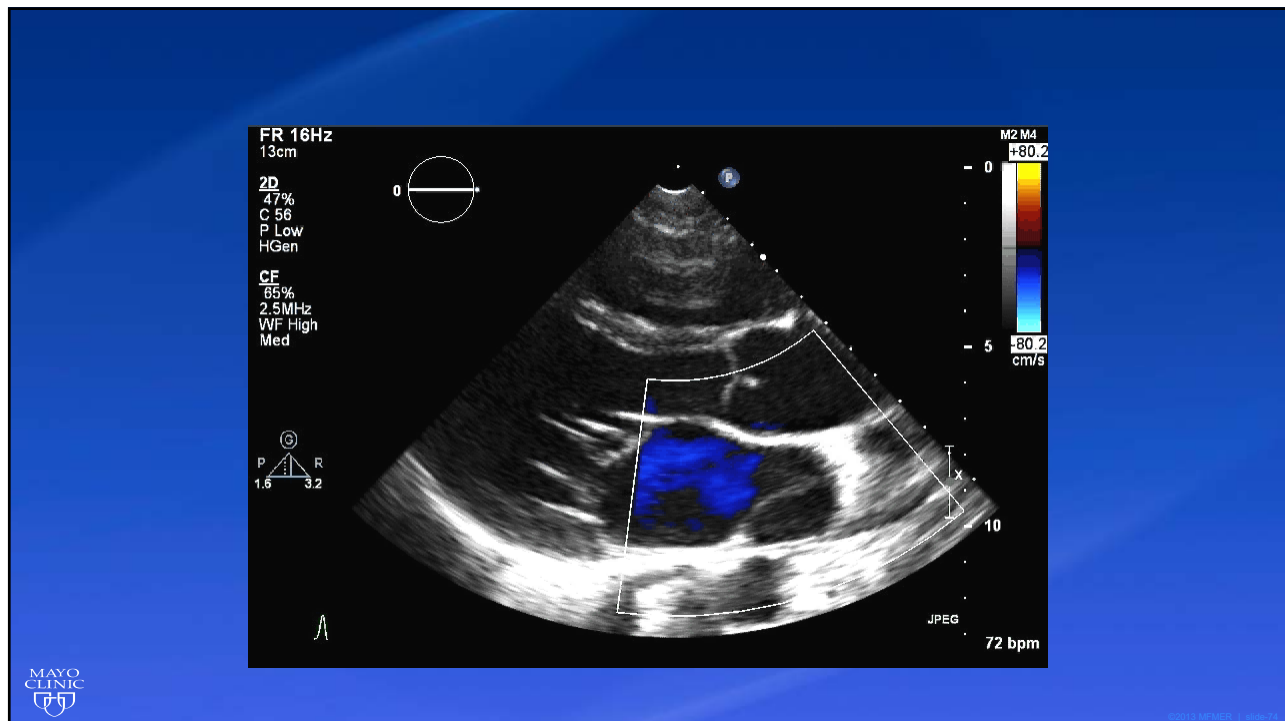
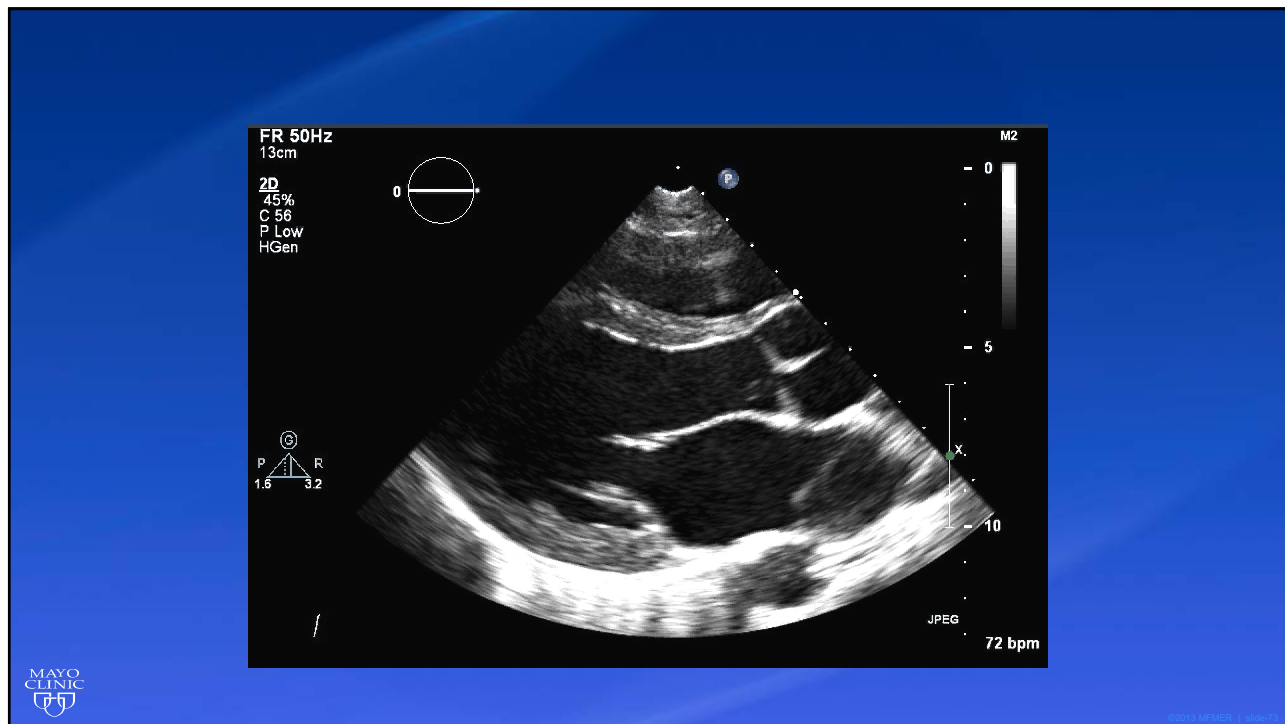


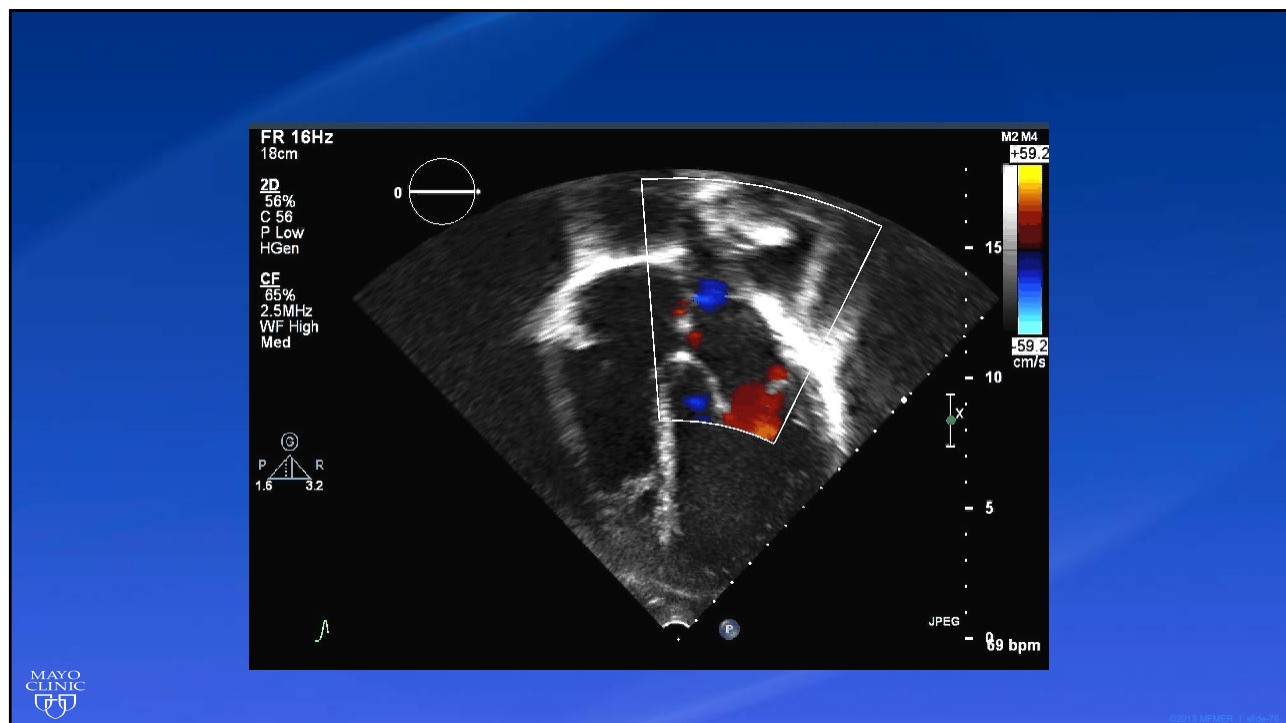
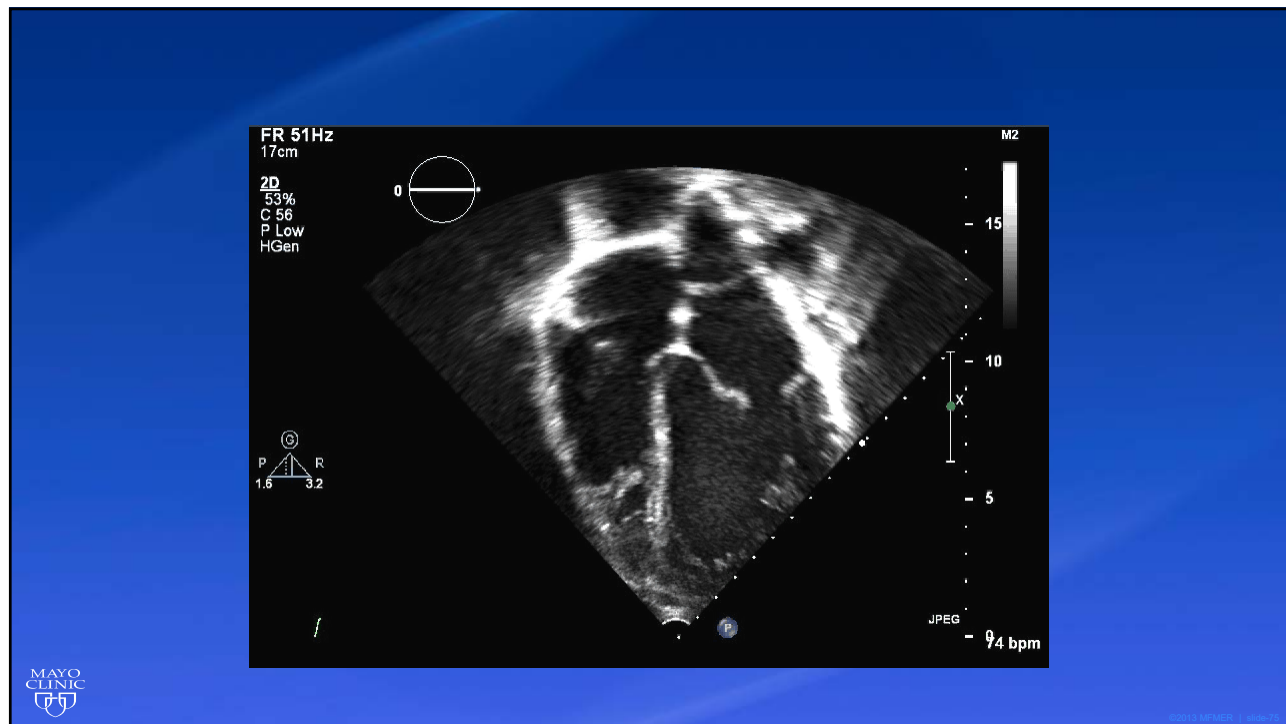


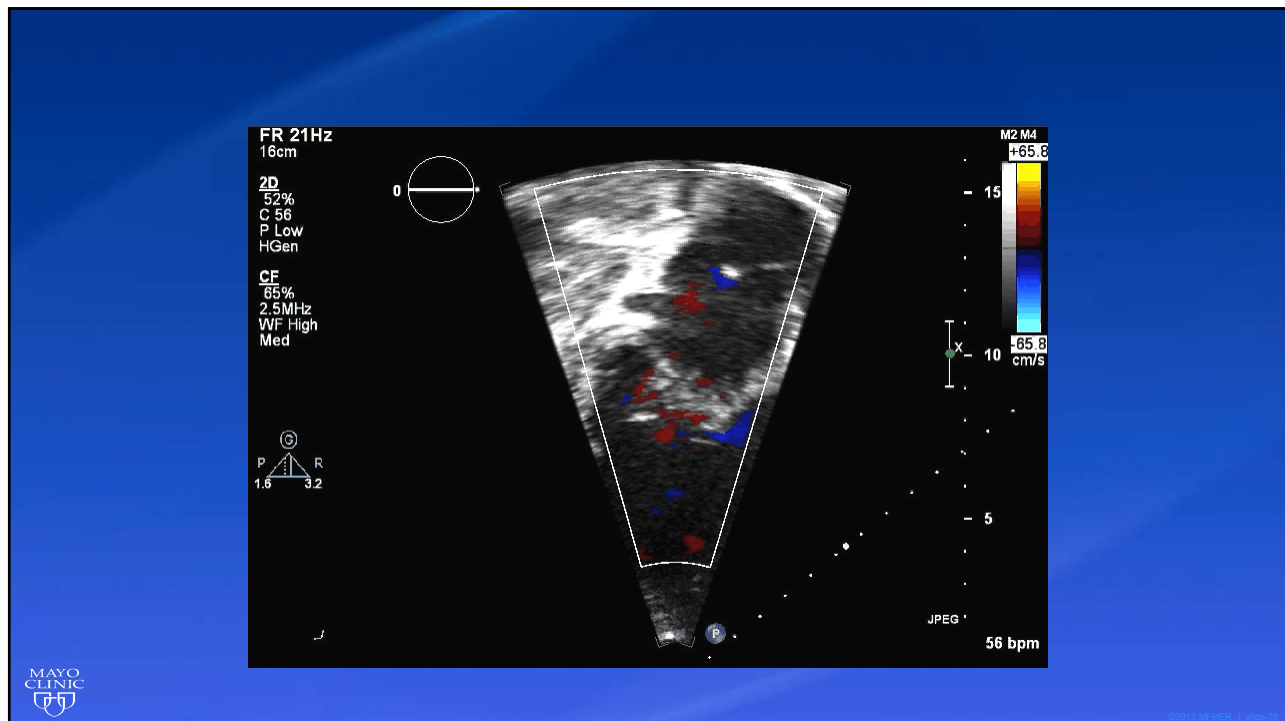
## Cor Triatriatum

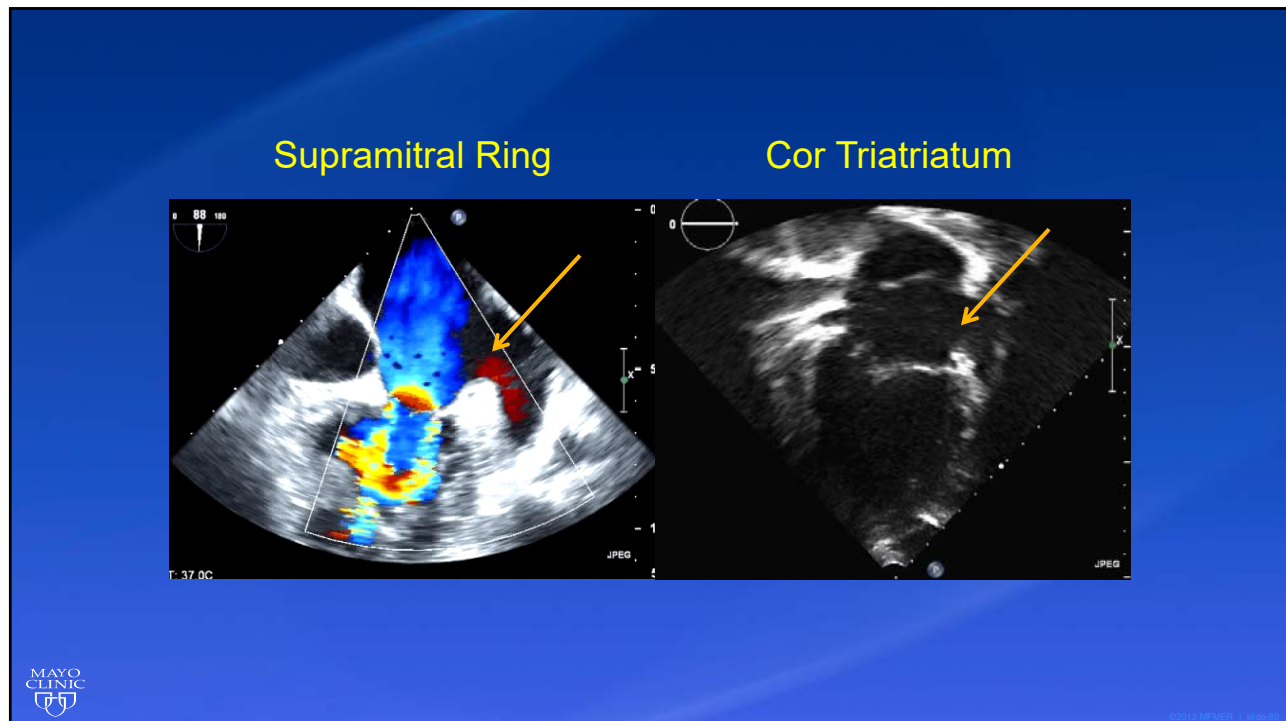
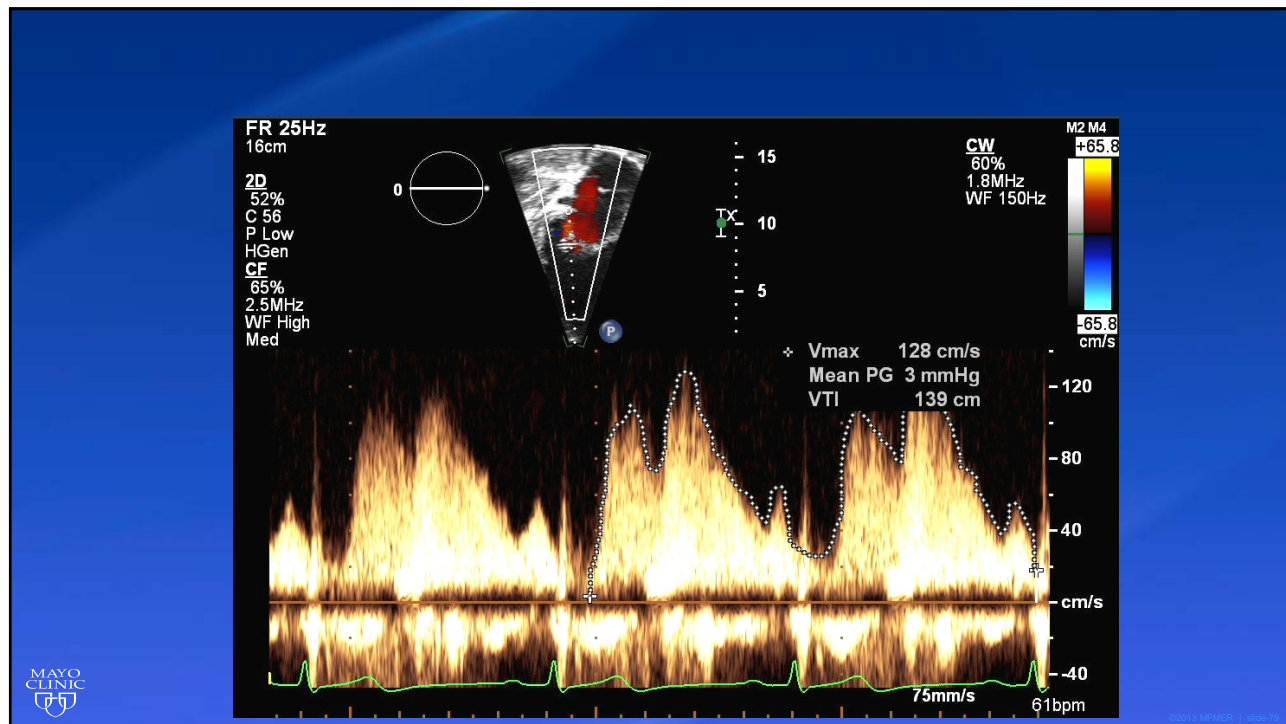
- Failure of proper embryologic development of the common pulmonary vein
- Mitral valve structure usually normal
- Fibromuscular membrane *proximal* to the left atrial appendage that divides the atrium into two parts









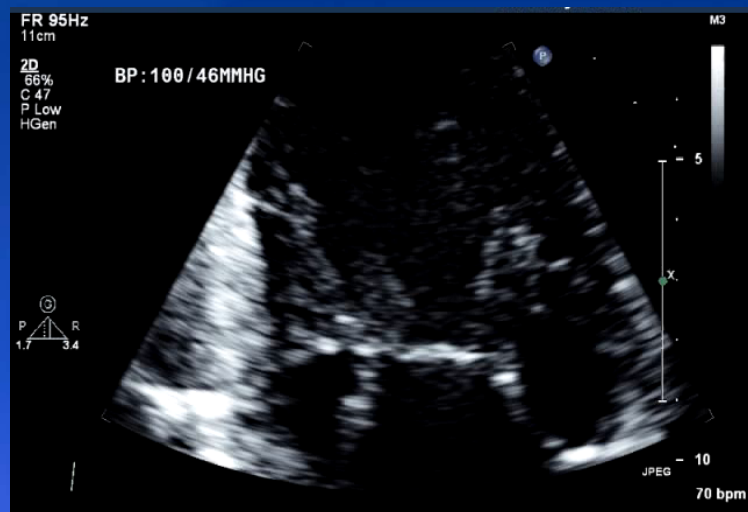


## Mitral Arcade

- Caused by an arrest in the developmental stage of the mitral valve before attenuation and lengthening of the chordae
- Chords are thickened, very short, or absent
- Fibrous bridge may join the two papillary muscles
- Results in both stenosis and regurgitation



©2013 Mayo Clinic. All rights reserved.



©2013 Mayo Clinic. All rights reserved.

